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Case Report

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Concomitant Fibrous Dysplasia and Aneurysmal Bone Cyst of the Skull Base

Case Report and Review of the Literature

Key Words
Aneurysmal bone cyst
Fibrous dysplasia
Skull base
Bone neoplasm
MRI

Abstract
The combination of fibrous dysplasia and aneurysmal bone cyst of the skull base is extremely rare yet it should be recognized, lest the rapid growth of the lesion and the radiological picture lead to the wrong diagnosis of a malignant process. We report on a 6½-year-old patient who suffered from concomitant fibrous dysplasia and aneurysmal bone cyst of the skull base. He is still disease-free 4 years postoperatively. The treatment of choice remains early surgical removal of the maximal amount of involved bone that can safely be achieved. There is no role for radiotherapy in the treatment of these lesions. A review of the literature is included.

Introduction

We report on a young boy affected with an extensive bony lesion of the lateral skull base. The lesion was resected and the patient remains disease-free 4 years postoperatively. Pathology revealed features of both fibrous dysplasia (FD) and aneurysmal bone cyst (ABC). The combination of these two pathologies in the skull is very rare. We reviewed the world literature and present recommendations as to the proper diagnosis and optimal management of such lesions.

Case Record

A 6½-year-old boy presented with a painless bulge over the right temporal area, enlarging slowly over the previous 2 years. The mass was solid, nontender and not movable, the overlying skin appeared normal. Hearing and vision were unaffected. A plain CT scan performed in May 1992 revealed an expansible bony lesion measuring 4 cm in its largest diameter, involving the floor of the right middle fossa and the right temporal squama (fig. 1A). The lesion was mainly cystic with a small solid component involving the greater sphenoid wing. In January 1994, a repeat enhanced CT scan revealed a mass consisting of a solid and a cystic component. The solid portion was embedded with small cysts and involved mainly the lateral orbital wall and the floor of the middle cranial fossa, extending inferiorly into the infratemporal fossa (fig. 1B, C). It enhanced homogeneously after contrast injection. The cystic component protruded into the middle cranial fossa displacing the right temporal lobe superiorly and posteriorly and was composed solely of multiple large cysts (fig. 1D). The bony cortices were severely expanded and thinned but not eroded. Medially, the cysts abutted the cavernous sinus without invading it. This component showed only peripheral enhancement after contrast administration. No adjacent brain edema was noted. The list of differential diagnosis at the time consisted of: ABC or a chondromatous tumor (benign or malignant). Two incisional biopsies were performed. The first (January 27, 1994) was inconclusive, the second (February 18, 1994) was read as meningioma.

The patient was referred to us in June 1994. The mass was nontender and had enlarged to a size of 9 × 6 cm. The overlying skin had become extremely stretched, glistening and erythematous (fig. 2A).

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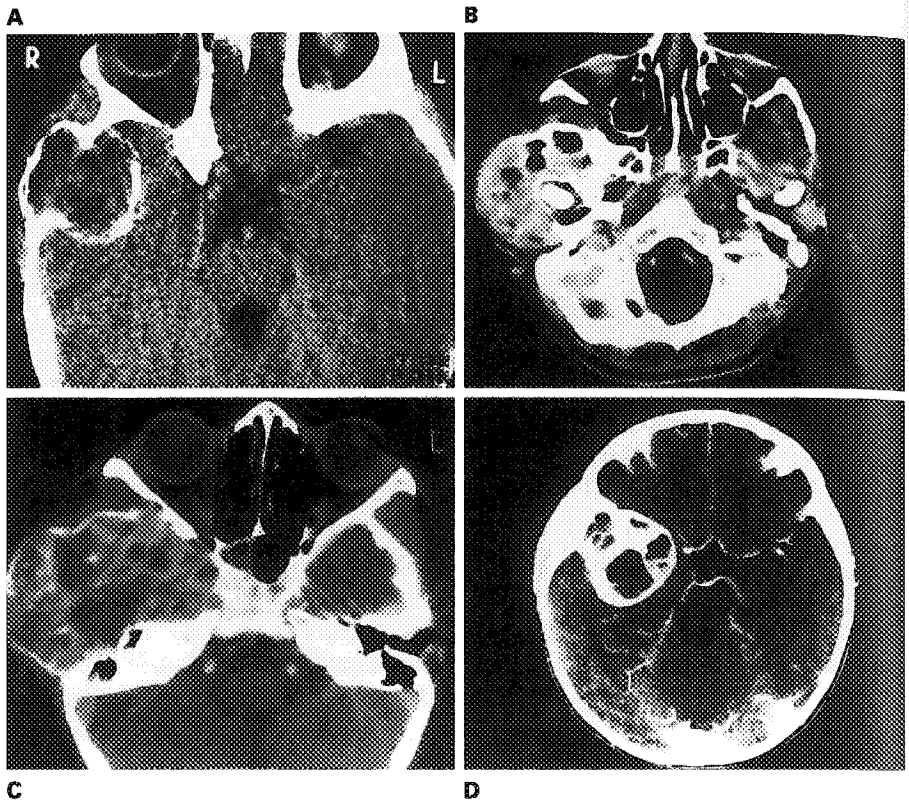


Fig. 1. Preoperative CT scans. **A** (Plain CT, 1992). Note the cystic ballooning of the right temporal squama. **B** (Enhanced CT, 1994, soft tissue window). Note the homogeneous enhancement of the subtemporal component. **C** (Enhanced CT, 1994). Bone window imaging of the floor of the right middle fossa. **D** (Enhanced CT, 1994, soft tissue window) note the cystic component displacing the temporal lobe posteriorly and superiorly.

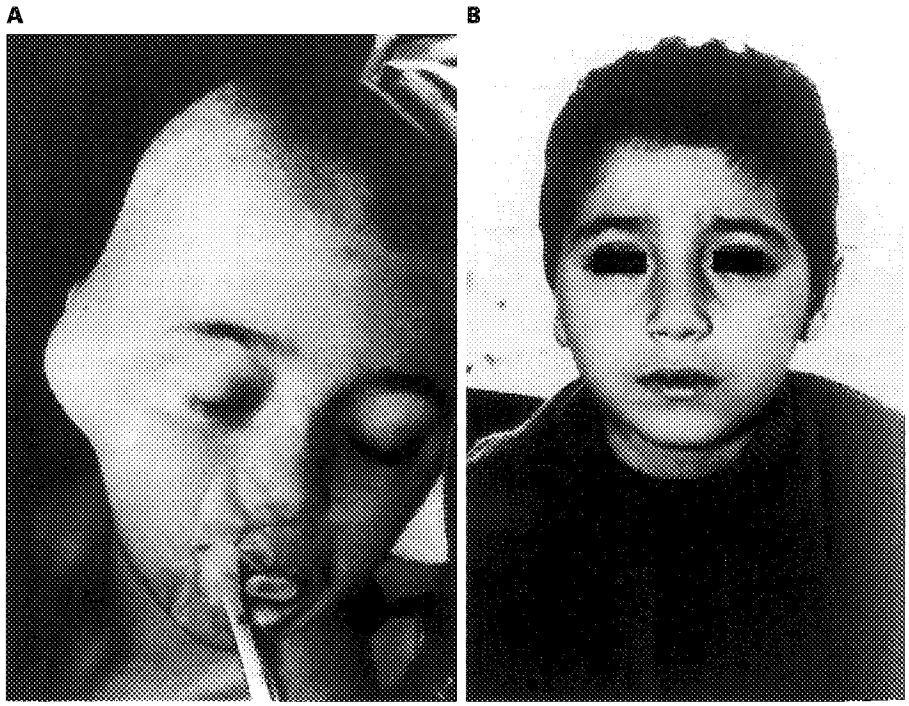


Fig. 2. The patient preoperatively (**A**) and 14 months postoperatively (**B**).

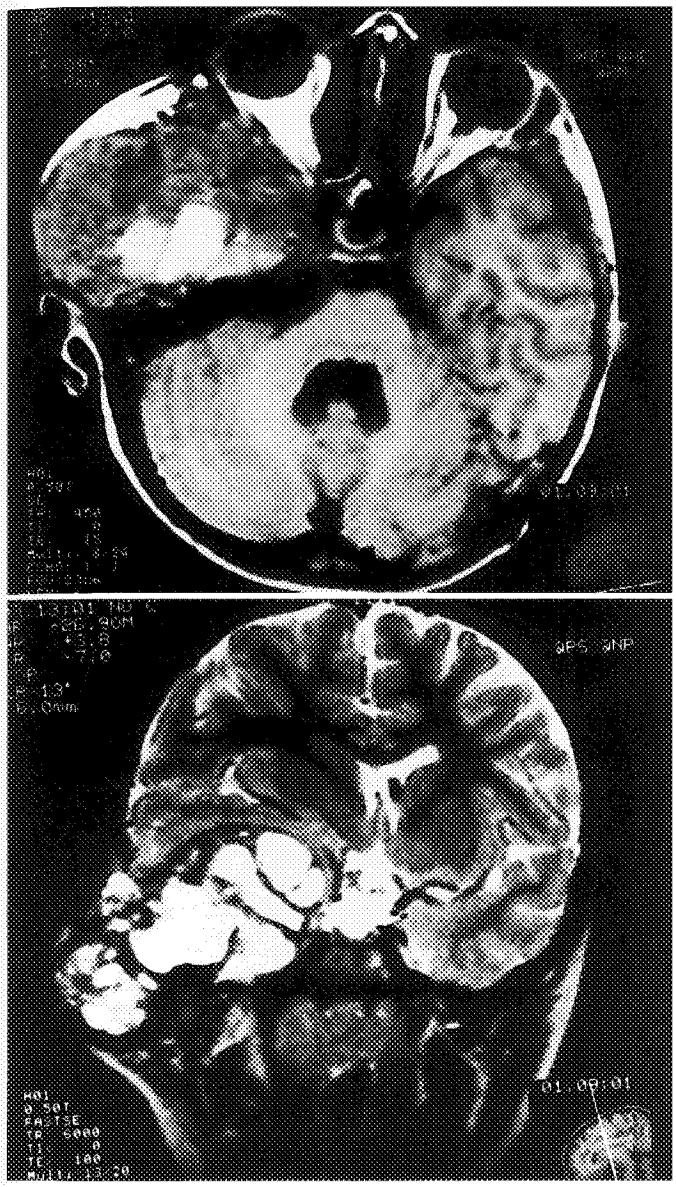


Fig. 3. Preoperative MRI. **A** Axial T1W. **B** Coronal T2W images revealing both components. Note the hyperintensity of some cysts seen in **A**.

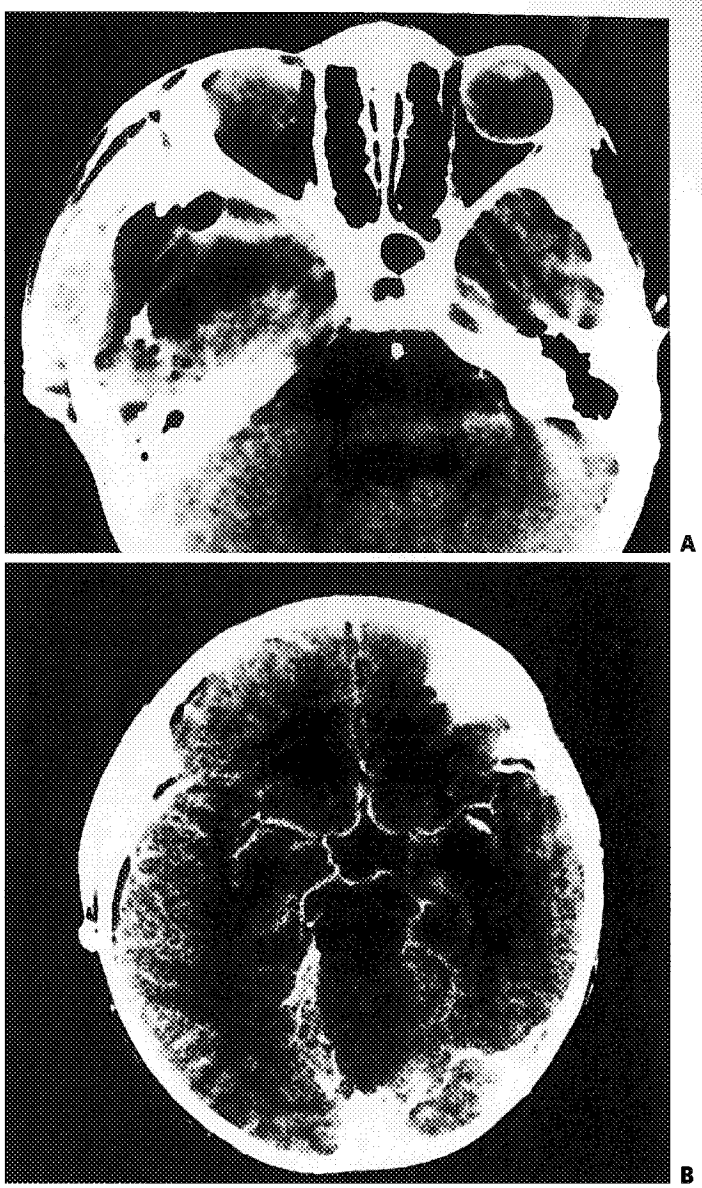
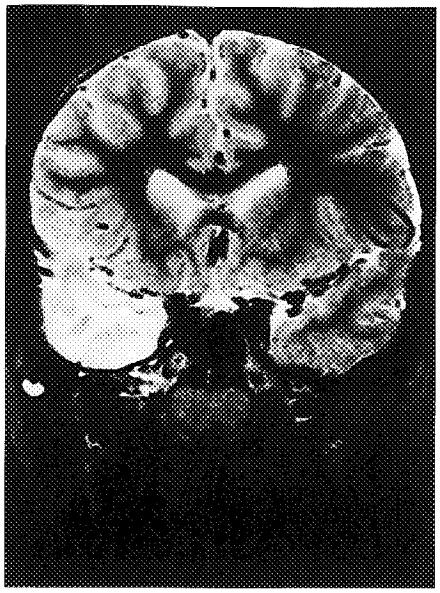


Fig. 4. Postoperative CT scan reveals complete resection of both the solid (**A**) and the cystic (**B**) components.

He complained of decreased hearing and tinnitus in the right ear of 2 weeks' duration. The neurological examination was otherwise normal. No café-au-lait spots were detected. An MRI revealed again the two previously described components. The solid portion was isointense with normal grey matter on T1-weighted (T1W) images (fig. 3A) and isointense to slightly hyperintense on T2-weighted (T2W) and proton density images. The cystic component was uniformly hyperintense on T2W images (fig. 3B). Three cysts revealed fluid-fluid levels with high signal on T1W, T2W and proton density images denoting a hemorrhagic content. Magnetic resonance angiography did not reveal any abnormal vascularization.

Surgery was performed on July 11, 1994. The patient was placed in the supine position with the right shoulder elevated and the head turned to the left. The mass was thus placed at the highest point in the surgical field. After achieving proximal control of the right cervical carotid, a question mark incision was performed from the pretragal to the frontal areas. The zygoma was severly thinned out and was excised. A frontotemporal craniotomy through uninvolved bone was performed to allow wider access to the skull base. The tumor was not adherent to the overlying soft tissues. The cortical bone was mainly preserved even though it was eroded in a few places. The tumor filled the infratemporal fossa and extended intracranially through the floor



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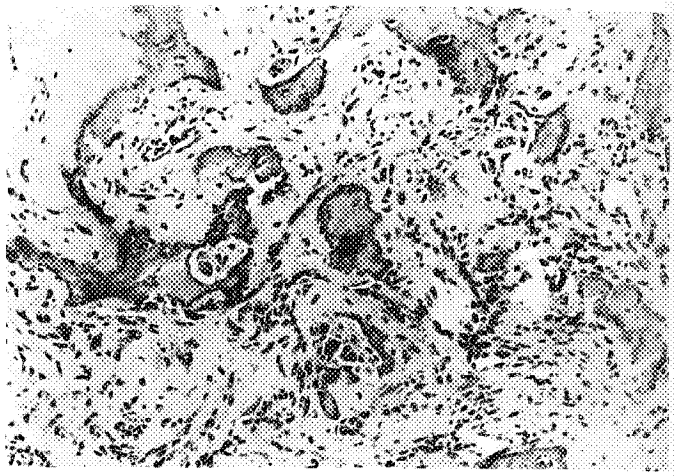

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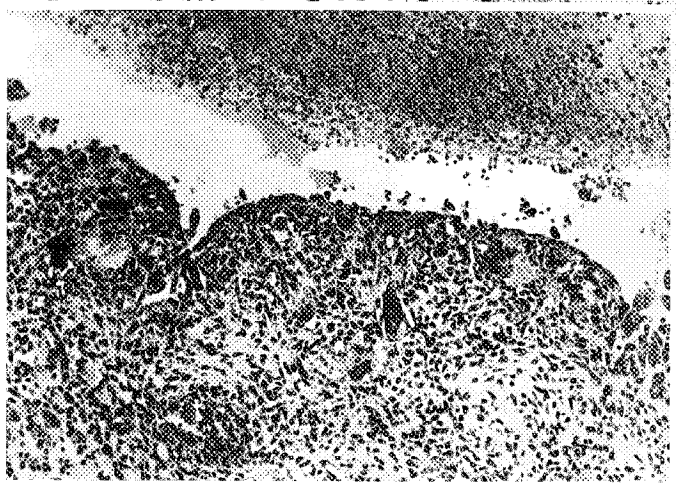

6B

Fig. 5. MRI performed 14 months after surgery shows no recurrence. Note the residual gliosis in the temporal lobe (coronal, T2W image).

Fig. 6. A Features of fibrous dysplasia showing mainly a fibrous matrix and immature bony trabeculae mimicking Chinese letters. The absence of osteoblastic rimming should be noted. H & E. × 250. **B** Features of aneurysmal bone cyst showing mainly a hemorrhagic cavity (superiorly) lined by a fibrous wall (inferiorly). Note the paucity of endothelial cells. Few giant cells and fibroblasts are seen. H & E. × 250.

of the middle fossa elevating the temporal lobe and displacing it posteriorly and superiorly. Horizontally, the lesion extended from the lateral orbital wall and cavernous sinus anteriorly to the petrous bone posteriorly. The two components of the tumor were identified. The solid component was vascular and gritty. It was removed in a piecemeal fashion. The cystic component consisted of markedly thinned cortical bone surrounding membranous cysts containing machinery oil fluid. The solid and cystic components were intermingled in areas. The dura was not adherent to the tumor and no dural reaction was noted. The involved portion of the sphenoid bone was resected. The carotid canal was dehiscent and the horizontal portion of the carotid was identified. The Eustachian canal was entered and was obliterated with bone wax. All the affected bone was excised and the bony edges were thoroughly drilled until normal bone was encountered. The craniotomy flap was replaced and secured. The galea and skin were closed in the usual fashion. Postoperatively, the patient fared very well. He was discharged on postoperative day 7. A CT scan performed directly postoperatively (July 15, 1994) revealed complete resection of the tumor (fig. 4). However the lateral orbital wall was till thickened. Eight months later, an MRI confirmed the complete gross resection of the lesion. The floor of the middle cranial fossa and the anterolateral portion of the petrous bone showed an abnormal signal characterized by a low T1W and a high T2W intensity. These abnormalities most probably represent postoperative changes. The temporal lobe that was markedly compressed preoperatively was back to its normal position. A repeat MRI, 14 months postoperatively, showed no evidence of recurrence (fig. 5). Cosmetic and functional results are still extremely satisfactory, 4 years after operation, with no clinical evidence of recurrence (fig. 2B). It is to be noted that, had the patient been operated at the time of the initial presentation in May 1992, surgery would have been easier and less extensive.

The two different components were noted on histopathology. The tissue removed from the solid component was composed of a fibrous background intermixed with immature bone trabeculae showing the classical 'Chinese letter' pattern seen in FD (fig. 6A). There was a noticeable absence of osteoblasts and osteoclasts. Tissue removed from the cystic component showed thin irregular membranes containing old hemorrhages. No endothelial cell lining or mitotic figures were noted. Areas of giant cells, immature bone formation rimmed by osteoclasts and numerous reactive, regular spindle cells were seen (fig. 6B). Thus the lesion had features of FD in the solid areas and of ABC in the cystic areas.

Discussion

FD is an idiopathic condition in which normal bone is altered by abnormal fibroosseous tissue causing distortion and overgrowth of the affected bone [1]. The areas of fibrous tissue are interwoven with newly formed bone trabeculae that vary in size and shape, the Chinese character trabeculae [1]. FD has been classified into three categories: (1) monostotic FD where only one bone is involved (70%); (2) polyostotic FD where multiple bones are involved (30%), and (3) McCune-Albright syndrome where FD is associated with café-au-lait spots and multiple endocrinopathies (rare) [2].

FD is usually diagnosed in the first three decades of life [1, 3]. The lesions tend to stabilize after puberty [4]. However this is not invariably the case [2]. The growth of FD may be affected by other endocrine factors such as pregnancy [5, 6]. It is usually a benign process, even though malignant degeneration has been reported in 0.5% of monostotic FD and in 4% of patients suffering from McCune-Albright syndrome [7]. Forty-six percent of cases that developed malignancy had received prior radiotherapy [7]. Some authors have stated that FD of the skull respects bony sutures, this is not always the case as evidenced by our patient [8]. Thus differentiation between monostotic and polyostotic FD in craniofacial cases is sometimes very difficult [9]. Craniofacial FD usually presents as a painless mass [9]. Symptoms are secondary to mass effect (e.g. proptosis, encroachment on middle ear structures, obstruction of ostia leading to epiphora or sinusitis, or involvement of the foramina of the skull base) [8–12]. The three most common radiological appearances are the pagetoid, sclerotic and cystic patterns [9, 10]. The latter pattern is seen in our patient. ABC are rare lesions. The great majority of patients are younger than 20 years [3, 13, 14]. On histopathology, ABC appear as blood-filled, cavernous spaces with a scarcity of endothelial cells. The cysts are separated by septae composed of spindle-celled fibrous tissue containing multinucleated giant cells and possibly osteoid tissue [1, 3, 15]. The surrounding bony cortex is 'blown out' into a very thin bony shell [14]. Both skull tables may be involved [16]. In a variant of ABC termed 'solid aneurysmal bone cyst', reticulated chondroid-like material is found whereas the typical cystlike spaces are absent [17, 18]. Plain X-rays of ABC reveal either a single or multiple 'blown out' cyst [14]. CT scan reveals an expansible bony cyst that may be multiloculated and may enhance peripherally [13, 16, 19]. Fluid-fluid levels may be seen to a better advantage on MRI [14]. The T1W signal of some cysts may be hyperintense, presumably secondary to methemoglobin [14]. ABC occur most commonly in the vertebrae and the long bones [3]. A recent review of the literature found 60 cases of ABC affecting the skull [13]. They usually present as an enlarging mass [13, 16, 19]. Symptoms may arise secondary to mass effect (headache, proptosis, seizures) [16]. More catastrophic presentations such as intracerebral hemorrhages have been reported [20]. The natural history of ABC is described in four stages: initial, growth, stabilization and healing stages [14]. With the possibility of ABC being grafted on a malignant lesion, it is difficult to evaluate the rate of malignant degeneration of a 'pure' ABC. The malignancy transformation rate has been quoted as 3% [3]. Treatment of skull ABC has included surgical extirpation, curettage, aspiration, radiotherapy and embolization [2, 13, 16, 21]. Calcitonin and estrogen therapies have been used in noncranial ABC [13, 19].

Theories for the etiology of ABCs invoke a preexisting trauma, a vascular etiology, or the possibility that ABCs may be just an epiphenomenon to another primary pathology [19]. Some authors stress that a preexisting lesion should always be sought when dealing with ABC, the latter being commonly associated with chondroblastomas and giant cell tumors [14, 15].

The association of ABC and FD is exceedingly rare. In a report of 57 ABCs associated to other osseous lesions, Levy et al. [22] did not find a single case of FD. In a review of 42 cases of FD Martinez et al. [15] found only 1 case of ABC, 9 other cases showed nonspecific cystic changes. A few detailed cases of extracranial concomitant ABC and FD (FD/ABC) have been reported [6, 23, 24]. All the cases of skull FD/ABC reported in the western literature have been reviewed and are summarized in table 1 [24–26]. It is to be noted that at least 3 of the cases of FD/ABC were characterized by rapid enlargement of a previously dormant lesion as in our case. The rapid enlargement should not be misdiagnosed as malignant degeneration of FD, it should be recognized as aneurysmal bone cyst transformation and dealt with accordingly. The reported cases are too few to delineate the exact natural history of such a lesion. Graf et al. [27] in 1980 reported a spontaneous recurrent hemorrhage as a complication of skull FD. The lesion was treated by repeated aspirations. We believe that this patient may have suffered from FD/ABC of the skull because of the typical presentation and the repeated episode of intralesional bleeding. Based on this report, there may be a place for repeated aspirations of the cyst in some cases. The preferred treatment, however, for symptomatic FD/ABC of the skull is early surgical excision of the largest amount of

Table 1. Reported cases of concomitant fibrous dysplasia and aneurysmal bone cyst of the skull

Author/ year	Sex/ age	Location/ size	Symptoms	Radiology	Treatment	Follow-up	Remark
Branch et al., 1986 [25]	F 9	Left parietal & fronto-temporal 4 × 5 cm	Cystic, tender mass (parietal); firm, nontender mass (frontotemporal)	SXR: increased opacity of both areas indicative of fibrous dysplasia; CT: large area of bone lysis with expansion of the diploic space in the parietal lesion	Surgical excision of the parietal lesion; no dural invasion	None	Left frontal lesion appeared 2 years prior to the parietal lesion; the left parietal lesion became tender and increased in size 1 month prior to presentation
Branch et al., 1986 [25]	M 19	Right parietal 6 × 8 cm	Painless mass	SXR and CT: cystic expansion of the skull	Surgical excision; no dural invasion	None	Patient was a known case of FD, followed for 15 years; the frontal lesions remained dormant; the parietal lesion increased in size over 4 weeks
Rappaport 1989 [26]	M 25	Left occipital 6 × 4 cm	Tender mass	SXR: Cystic expansion of diplöe, sclerotic margins; CT: pagetoid changes in the left orbital area; with intradiploic hypodense lesion	Surgical excision; no dural invasion	None	Patient was a known case of FD; lesion appeared after local trauma; the cystic lesion was punctured percutaneously prior to surgery but refilled within 10 min; the actual pathology did not reveal fibrodysplastic tissue
Wojno and McCarthy 1994 [24]	F 14	Right temporal 5 cm	Painless mass	CT: nonhomogeneous cystic mass	Surgical excision	Two years disease-free	Preoperative diagnosis of low-grade malignancy
Wojno and McCarthy 1994 [24]	M 40	Left frontal 5 cm	Expanding mass	CT: diffuse thickening of the calvarium, left frontal cyst with septation	Surgical excision	None	Mass appeared after head trauma
Haddad 1996	M 6.5	Left skull base	Expanding mass	CT: nonhomogeneous cystic mass; MRI: two components, solid and cystic	Surgical excision	Four years disease-free	

SXR: Skull X-ray; CT: computed tomography; MRI: magnetic resonance imaging.

diseased bone that can be safely removed. There is no evidence that postoperative radiotherapy is useful. On the contrary, it may lead to malignant transformation [25].

Conclusion

Even though FD may present as a cystic variant and ABC as a solid variant, we believe that our patient did not belong to either preceding categories but harbored two distinct pathologies, namely FD and ABC. Our conviction stems from the clear characteristic radiological and pathological pattern of each component. The clinical evolution and the radiological findings of such a double lesion do not indicate malignancy. Early surgery remains the preferred method of treatment. The surgeon should aim at removing the greatest amount of diseased bone without jeopardizing the functional outcome of the patient. Functional and cosmetic results of surgery can be very rewarding.

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